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Case Series

Malignant tumors of the small intestine: A case series

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Abstract

Small bowel malignancies are rare, yet their incidence has been rising in the last decade. The main histological types are adenocarcinomas, neuroendocrine and stromal tumors, sarcomas, and lymphomas. The clinical presentation is often nonspecific, making it a challenging diagnosis that results in delayed treatment. We report a case series of four patients who present with non-specific gastrointestinal symptoms and a subsequent diagnosis of a small bowel malignancy. While advanced endoscopic techniques have improved our ability to identify these uncommon tumors, in our case series, the definitive diagnosis was delayed due to the ambiguous presentation. Larger studies are warranted for better diagnosis and treatment of small bowel malignancies.

Introduction

Small bowel neoplasms constitute less than 3% of all gastrointestinal malignancies and 0.6% of all cancers in the United States [1-3]. While rare, their incidence has steadily increased in the last 20 years [3]. The most common histologic subtypes are adenocarcinoma and neuroendocrine tumors, each accounting for approximately 40%. Stromal tumors, sarcomas, and lymphomas comprise the remaining 20% [4-8]. Symptoms are non-specific and include abdominal pain, weight loss, nausea, vomiting, obstruction, and occult bleeding [9,10]. Clinical signs are vague, the physical exam is frequently unremarkable, and visualization on radiological imaging is limited by motion artifacts, making it a challenging diagnosis [2,11]. Endoscopic techniques, video capsule endoscopy, and push enteroscopy have improved our ability to identify these uncommon tumors. However, unclear clinical signs and symptoms can lead to late diagnosis and treatment. We present a case series of four patients with vague clinical presentations who underwent extensive workup with advanced imaging modalities and were eventually diagnosed with a small bowel malignancy.

Case report

Patient 1

A 43-year-old male with a past medical history of sarcoidosis presented with three months of worsening periumbilical pain and a 14-kilogram weight loss. His symptoms were initially attributed to pancolitis, treated with oral antibiotics. Prior to infectious workup, serology for Inflammatory Bowel Disease (IBD), Esophagogastroduodenoscopy (EGD), and colonoscopy were unrevealing. Video Capsule Endoscopy (VCE) demonstrated localized inflammation in the ileum, however, the capsule was unable to pass beyond this point (Figure 1). The physical examination, laboratory investigation, and chest radiograph were unremarkable. CT abdomen and pelvis revealed a partial small bowel obstruction. Small bowel enteroscopy demonstrated nonspecific inflammation of the ileum.

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CT enterography disclosed the presence of a stricture in the mid-ileum (Figure 2). Given the unclear etiology and persistent symptoms, three months following the initial presentation, small bowel resection with side-to-side anastomosis was performed. Operative findings included an ileal stricture but otherwise normal bowel. Pathology revealed diffuse large B-cell lymphoma (DLBCL) of the small intestine (Figure 3) and the patient was treated with R-CHOP chemotherapy [12].

Patient 2

A 55-year-old female with a past medical history of Lynch syndrome and a family history of colon cancer presented with abdominal pain, nausea, vomiting, and a 2.3-kilogram unintentional weight loss for one month. The physical examination and laboratory investigation, including IBD serology, were unremarkable. Magnetic resonance enterography showed a 6-centimeter (cm) proximal ileal segment with evidence of irregular concentric wall thickening. Small bowel enteroscopy revealed a white nodular ileal mucosa with areas of ulceration in the mid-ileum (Figure 4). Biopsies demonstrated low-grade follicular lymphoma, four months after initial presentation.

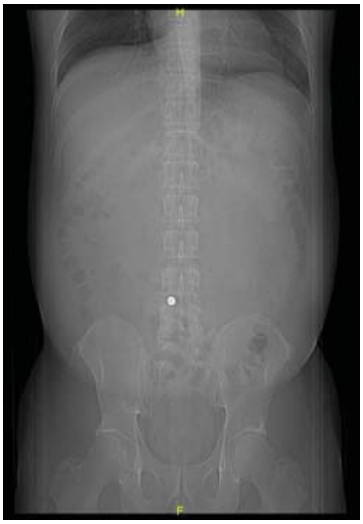

Figure 1: Coronal image of computed tomography in scout view showing lodged endoscopic capsule.

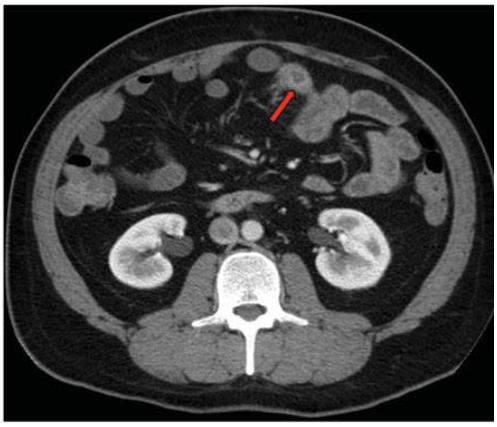

Figure 2: Computed tomography enterography showing mild hyperenhancement in the narrowed region (red arrow).

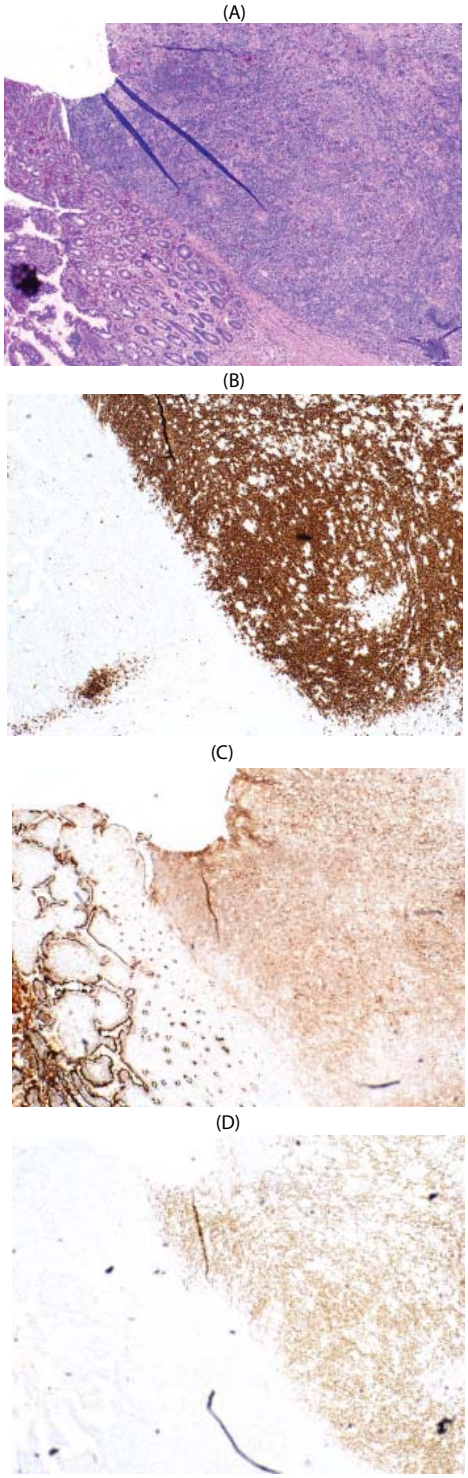

Figure 3: (A) Lymphoma involving the bowel wall. (B) CD20 positive immunostain. (C) CD10 positive immunostain. (D) Large B-Cell, Ki-67 stain (more than 70%).

Patient 3

A 68-year-old female with a past medical history of breast cancer presented with one month of abdominal pain, bloating, and diarrhea. The physical examination and laboratory investigation work were unremarkable. Upper endoscopy revealed an antrum nodule with regenerative changes and a hyperplastic duodenal bulb nodule with preserved villous architecture. Initial pathology revealed reactive gastropathy in

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the antrum and a benign hyperplastic/inflammatory polyp in the duodenum. Further evaluation with EGD and Endoscopic Ultrasound (EUS) indicated a 10 millimeter (mm) by 12 mm intramural lesion in the antrum of the stomach that was most consistent with a lipoma. Additionally, a hypoechoic 13 mm x 12 mm round mass in the duodenal bulb was seen confined to the mucosa (Figure 5). Endoscopic mucosal resection of the duodenal lesion was performed. A well-differentiated neuroendocrine tumor, low-grade World Health Organization (WHO) Grade 1 and 3, with tumor involvement of the muscularis mucosa was confirmed on biopsy six months following initial presentation.

Patient 4

A 53-year-old male without significant medical history presented following one month of vomiting and epigastric pain. A gastric emptying study showed 60% food residual with a prolonged gastric emptying half-time. Laboratory investigation, EGD, colonoscopy, and CT abdomen and pelvis were unremarkable. His symptoms were initially attributed to gastroparesis and was treated with domperidone. He presented six months after the initial presentation with a 23-kilogram weight loss and was treated for refractory gastroparesis with metoclopramide and erythromycin. He was scheduled to undergo a gastric peroral endoscopic myotomy procedure, however, EGD demonstrated 5 liters of fluid in a severely dilated duodenum suggesting an obstruction (Figure 6). Diagnostic laparoscopy revealed a nearly obstructing mass. A small bowel resection was performed, and moderately differentiated, invasive adenocarcinoma, invading through muscularis propria into nonperitonealized perimuscular tissue (mesentery and retroperitoneum) without serosal penetration, was confirmed on pathology (Figure 7).

Discussion

Small bowel cancer is a rare disease; however, the incidence is on the rise, with an estimated 12,070 new cases and 2,070 deaths in the United States in 2023 [3]. Adenocarcinoma and neuroendocrine tumors are the most common histological subtypes of small bowel malignancies.

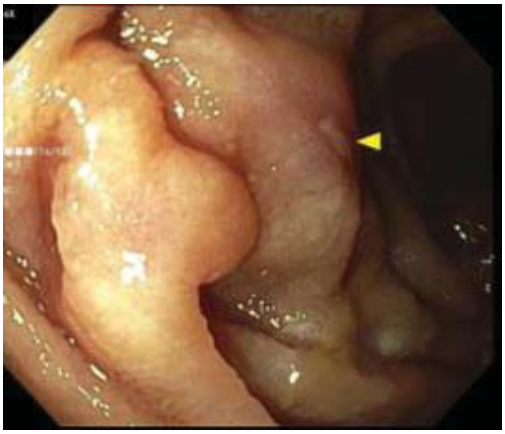

Figure 4: Small bowel enteroscopy demonstrating a white ulcerated nodular mucosa in the ileum.

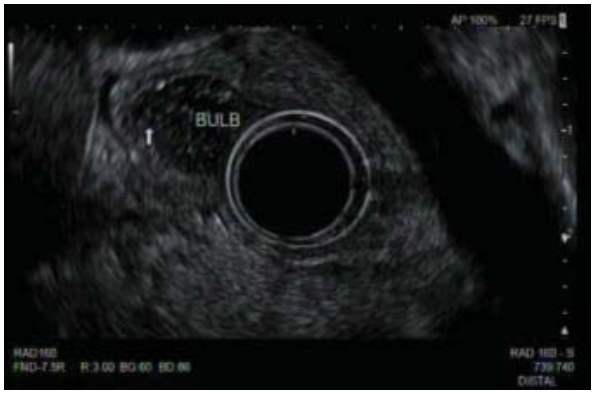

Figure 5: Endoscopic ultrasound revealing a hypoechoic lobulated mass in the duodenal bulb. The mass measured approximately 13 mm by 12 mm in maximal cross-sectional diameter. The lesion extended from the mucosa to the submucosa.

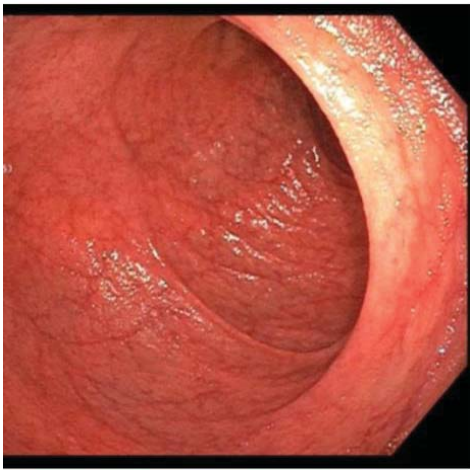

Figure 6: Esophagogastroduodenoscopy showing severe dilation of the duodenum.

Primary lymphoma of the gastrointestinal tract is especially rare and comprises 1% - 4% of all gastrointestinal malignancies [13]. The ileocecal region is one of the most involved areas for primary intestinal lymphoma and thus, it can mimic IBD and other colonic etiologies further delaying treatment due to its initial ambiguity. Typically, on radiographic imaging, small bowel lymphoma can present as a polypoid mass, multiple nodules, infiltrative form, an extraluminal mass, mucosal thickening, or in the form of strictures as seen in our patient with DLBCL (figure 2) [13]. However, CT imaging has a low sensitivity and specificity for detecting small bowel lymphomas. Thus, endoscopic evaluation can aid in the diagnosis of these tumors. In Figure 4, a white nodular ileal mucosa is seen in our patient diagnosed with follicular lymphoma on Small Bowel Enteroscopy (SBE), which is consistent with the typical findings of follicular lymphoma seen on endoscopy that include whitish polyps and white aggregates with or without ulceration of the mucosal layer [14-16]. Furthermore, EUS has enhanced our ability to visualize lesions of the gastrointestinal tract. As seen in Figure 5, a hyperechoic duodenal bulb lesion was identified and subsequently diagnosed as a neuroendocrine tumor. While several studies evaluated the role of EUS in detecting pancreatic neuroendocrine tumors, specific characteristics regarding lesions of the small bowel have yet to be established

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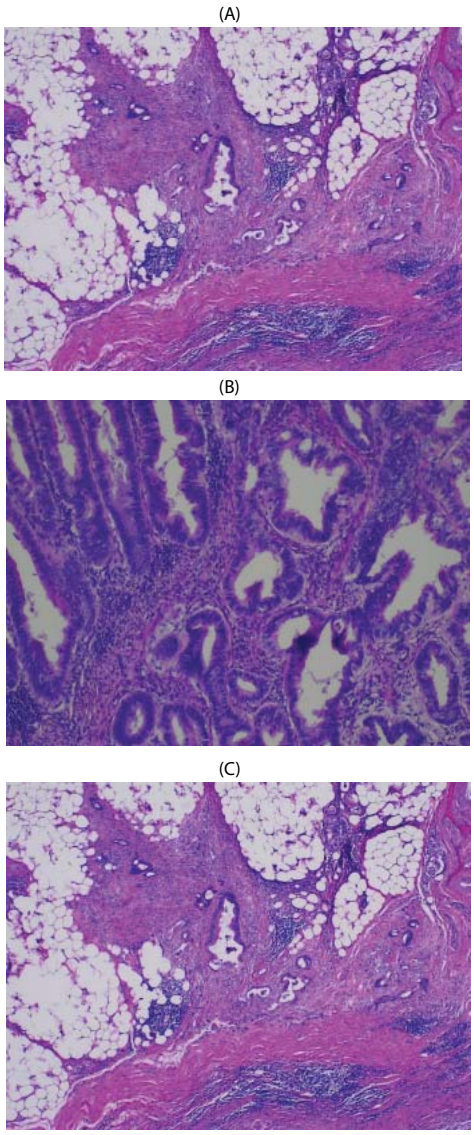

Figure 7: (A) Invasive adenocarcinoma invading into the pericolonic fat (adipose tissue), low power view. (B) Invasion into the submucosa and muscularis propria. (C) Invasive adenocarcinoma invading the pericolonic fat, high power view.

[17]. Given the rise in small bowel tumors, further studies are warranted to investigate the role of EUS in diagnoses of these malignancies. Additionally, intestinal ultrasound has been shown to accurately detect disease activity in the small bowel in patients with Crohn's disease. However, this inexpensive and non-invasive imaging modality has yet to be described for the specific detection of small bowel tumors [18,19].

Small bowel tumors are difficult to identify and there are no established guidelines on an initial testing strategy for diagnosis. We propose the following diagnostic approach for patients presenting with symptoms of intestinal disease such as abdominal pain, gastrointestinal bleeding, symptoms of small bowel obstruction with nausea and vomiting, weight loss or bowel perforation and there is a concern for a small bowel malignancy. Initial testing should include a non-invasive modality, abdominal imaging, either with CT or MRI to evaluate for any lesions. If no lesions are identified, but a high clinical suspicion remains, endoscopic evaluation may be performed to evaluate for a tumor and tissue biopsy if possible. Choice of endoscopic evaluation includes esophagogastroduodenoscopy, push enteroscopy, device-assisted endoscopy, illeocolonoscopy, and VCE. Modality should be chosen based on the individual patient's presenting symptoms. For example, VCE should be avoided in patients presenting with signs and symptoms of a bowel obstruction [20]. While several of these modalities were shown to assist in the diagnosis of localized small bowel adenocarcinoma, no single modality proved adequate for definitive diagnosis [21]. If no lesion was identified and there remains a high level of suspicion for a small bowel tumor, further imaging may be considered with CT enterography, fluorodeoxyglucose-positron emission tomography/CT, or somatostatin receptor-based imaging if there is a concern for a neuroendocrine tumor [22-24]. If the workup is nonconclusive, surgical evaluation may be considered.

Conclusion

In our series, presenting symptoms among all patients were consistent with non-specific gastrointestinal symptoms, an unremarkable physical examination, and normal laboratory investigation. While advanced endoscopic techniques have improved our ability to identify these uncommon tumors, in our case series, definitive diagnosis was delayed up to six months from the initial presentation due to the unclear etiology and treatments varied based on histologic subtype. The initial testing strategy in a patient suspected of having a small bowel tumor should begin with non-invasive imaging and subsequently endoscopic evaluation, choice of procedure chosen based on the patient's presenting symptoms. Larger and more powerful studies are needed to provide further insight into a more targeted diagnostic and treatment approach for improved clinical outcomes.

Data availability statement

The data that support the findings of this study are available from the corresponding author [SS], upon reasonable request. All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author [SS].

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